CUTANEOUS NEVI, COMMENTS AND COMMON MISCONCEPTIONS CONCERNING THEIR MANAGEMENT*

VICTOR H. WITTEN

Associate Clinical Professor of Dermatology and Syphilology, New York University Post-Graduate Medical School

AMONG the more common cutaneous lesions affecting man, those most likely to cause serious concern are certain of the pigmented and vascular nevi. Because of the importance of these particular nevi to individuals of all ages, regardless of sex or vocation, and because of the responsibility concerning these lesions that must be shouldered by the physician consulted, whether general practitioner or specialist, I have chosen to comment briefly on some of the more common misconceptions regarding the management of the more frequently encountered pigmented and vascular nevi. My discussion is based on recent publications prepared with Dr. Alfred W. Kopf[1] and Dr. Rudolf L. Baer[2], my colleagues in the Department of Dermatology and Syphilology of the New York University Post-Graduate Medical School.

I hope that, by discussing what I believe to be important "misconceptions", I shall be successful in dispelling some of the errors which have made their way into medical thinking, and as a result of which there may be errors in the management of these lesions.

Cutaneous nevi result from prenatal malformations, or, stated another way, are congenitally determined circumscribed new growths which may be present at birth, may appear shortly thereafter or any time later in life. The latter are referred to as delayed or tardive nevi.

PIGMENTED NEVI

Of the pigmented nevi, only those containing nevus cells, and which I shall refer to as nevus-cell nevi, will be discussed in that these are the ones that may prove of medical consequence. Nevus-cell nevi are of three types: 1) junction nevi; 2) compound nevi; 3) intradermal nevi.

Unfortunately, the gross features of nevus-cell nevi are not sufficiently typical to permit easy and accurate recognition. It is only

* From the Department of Dermatology and Syphilology, New York University Post-Graduate Medical School and the Skin and Cancer Unit of University Hospital, New York, N. Y.

through experience and a knowledge of the clinical characteristics of these lesions that any degree of proficiency can be achieved in establishing the correct diagnosis.

In general, *junction nevi* are usually tan to dark brown, and even blue to black, and the pigment is often mottled in distribution. The lesions are ordinarily flat to slightly elevated, smooth and usually without dark coarse hairs. They vary in size from 1 millimeter to several centimeters and may occur anywhere on the body, although they are most common on the face, extremities (including palms and soles) and upper chest and back.

Compound nevi may easily be confused with junction nevi because of the similarity of clinical characteristics, but unlike junction nevi, compound nevi are usually raised and may be either smooth or verrucous. At times they may resemble intradermal nevi although they are likely to be somewhat more darkly pigmented and flatter.

Intradermal nevi may appear skin colored or may have any gradation of pigment to dark brown; here, too, the pigment may appear mottled. The lesions are almost always distinctly elevated, being dome-shaped, polypoid or pedunculated and may or may not contain dark coarse hairs. They differ in size from 1-2 millimeters to 1 centimeter or larger, and may occur anywhere on the body surface, with a predilection for the face.

A word or two about the natural history and the histopathology of these cellular nevi should be both interesting and helpful in establishing the clinical diagnosis. It has been shown that in prepubertal children over 90 per cent of these nevi have junctional elements[3] whereas in adults only about 12 per cent contain such junctional activity[4]. This change in incidence and type of nevus can be explained by the series of events which take place with the growth and maturation of the individual. In the embryonic development of the fetus, the melanoblasts migrate from the neural crest to the basal layer of the epidermis and later are transformed into melanocytes and nevus cells. When these nevus cells come together in groups and are found mainly at the junction of the epidermis and dermis, that is, at the basal layer, the lesion is referred to as a junction nevus. In time, the nevus cells drop down from the basal layer to become located entirely within the dermis, and the lesion is designated an intradermal nevus. When the nevus cells are located at the junction of the epidermis and dermis as well as in the dermis, the lesion is known as a compound nevus.

The classification of Shaffer[5], which attempts to correlate the clinical and histopathologic findings of these lesions, may simplify and be of aid in making the correct diagnosis. According to Shaffer, plain, flat, pigmented nevi occurring before 30 years of age, and speckled, flat, slightly elevated and/or verrucoid nevi or those presenting an elevated central portion with a peripheral flat or slightly raised pigmented zone usually have junctional elements. On the other hand, flat, pigmented nevi in persons 30 years of age and older and those nevi which are polypoid, dome-shaped, sessile or pedunculated are, in most instances, intradermal nevi. At all ages, pigmented nevi on the palms, soles and anogenital region are, in all probability, junction nevi.

But why all the concern about the nevus-cell nevi? Why are they so important? Because it is only the junction and compound nevi which are known to have the proclivity to undergo malignant transformation into malignant melanomas. It should be pointed out, however, that according to estimates only one in a million nevi transforms to malignant melanoma[6]. If, as has been estimated, each individual has 20 nevi then only one in 50,000 persons will develop a malignant melanoma. Nevertheless, if malignant transformation from either the junction nevus or compound nevus can be avoided every precaution should be exerted. But there are certain misconceptions associated with this concern for these potentially malignant lesions. Among these are:

Misconception 1: If all nevus-cell nevi were excised prophylactically, the development of cutaneous malignant melanomas would be prevented.

Such an approach is not feasible because of 1) the impracticability of excising the conservative estimate of 3 billion 500 million nevi from persons in the United States alone, and 2) the fact that only 25-50 per cent of malignant melanomas of the skin would be prevented if excision of all the pigmented nevi could be accomplished, because a large percentage of malignant melanomas have their origin at sites clinically free of existing nevi[7].

Misconception 2: All nevus-cell nevi of the palms and soles should be excised.

Based on past figures which reported less than 1 per cent incidence of pigmented nevi on the palms and soles[8] and the therefore apparently high incidence of malignant melanoma developing from nevi at these sites, particularly the soles, excision of all such lesions has been widely

TABLE I.—INDICATIONS FOR THE TREATMENT OF NEVUS-CELL NEVI

Treatment Mandatory
1. Nevus-cell nevi which have undergone changes or have shown "activity" such as: sudden enlargement, bleeding, inflammation, alteration in color (either darker or lighter), ulceration, crusting, development of satellite pigmented areas, or those which become symptomatic (especially pruritic or painful).
2. Nevus-cell nevi showing "activity" during pregnancy[10].
3. Nevus-cell nevi which histologically show evidence of "unrest", e.g., precancerous junction nevi.
Treatment Generally Considered Advisable
1. Those closely related conditions with clinical features which suggest possible malignant transformation which is believed to take place in a reasonably high percentage of instances (e.g., melanotic freckle of Hutchinson or circumscribed precancerous melanosis[11]).
2. Benign juvenile melanomas[12].
3. Nevus-cell nevi in subungual areas.
Treatment Controversial
1. Nevus-cell nevi at sites of repeated trauma.
2. Nevus-cell nevi appearing beyond the second decade of life.
3. Nevus-cell nevi located on palms and particularly soles.
4. Nevus-cell nevi on genitalia and mucous membranes.

advocated. More recent figures, however, show that the incidence of pigmented lesions at these sites is closer to 25 per cent[9]. In view, then, of the frequency with which pigmented nevi appear on palms and soles, their removal from our entire population does not seem feasible. Furthermore, based on the comparatively low incidence of malignant melanomas at these sites, the excision of all such pigmented nevi does not seem warranted.

Misconception 3: One should never cut into a nevus, for example, to obtain a biopsy, because it will cause malignant transformation.

In our opinion this, too, is a misconception, for to our knowledge: 1) proof is lacking that cutting, or even traumatizing, a benign nevus-cell nevus has ever produced malignant degeneration; 2) we know of no histologically proven benign nevus-cell nevus which became malignant following biopsy; 3) malignant melanomas remain one of the less common malignant tumors (about 1 per cent) in spite of the fact that thousands upon thousands of nevi have been biopsied or destroyed; and 4) it is a prevalent opinion that malignancy already existed in those nevi for which the therapy administered was held to blame.

Of course, where malignant melanoma is strongly suspected on clinical grounds, diagnostic biopsy should not be performed without careful review of all the circumstances pertaining.

Misconception 4: All pigmented nevi should be removed.

We cannot agree with this precept for the various reasons already stated. The indications for the removal of pigmented nevi (including junction and compound nevus-cell nevi) are few. With the exception of treating them for cosmetic reasons, the indications for therapy are listed in Table I.

Treatment: When the decision has been made to remove a particular nevus, it is then necessary to choose a method which will be not only therapeutically correct but from which a cosmetically acceptable (though not always essential) result can be anticipated.

Junction and compound nevi, with few exceptions, should be surgically excised allowing but 1-2 millimeters of normal tissue at the periphery—in other words, the excision should be a conservative one. The excised specimen should be submitted for histopathologic examination by a pathologist specially qualified in tumors of the skin.

Intradermal nevi may be removed both safely and satisfactorily by a variety of accepted dermatologic procedures. The method will depend on the morphology and location of the lesion. Raised dome-shaped lesions, sessile, polypoid and pedunculated lesions may be cut off at the level of the skin surface with scalpel or scissors; any remaining tissue may be lightly electrodesiccated as required. Dark coarse hairs within a lesion should be removed *first* by electrolysis; sometimes the removal of the hairs alone will result in sufficient shrinking of the nevus to make it cosmetically acceptable.

For details concerning the recognition of these lesions and the various forms of therapy I would refer the reader to appropriate texts and articles on the subject.

Vascular Nevi

Only the three most common types of vascular nevi, that is, strawberry and cavernous hemangiomas and nevus flammeus (portwine mark) will be considered, and then only briefly.

Strawberry hemangiomas are superficial vascular proliferations which cause an elevated pink to red lesion, usually with an irregular

surface. With diascopy these soft tissues are readily compressible and the blood can usually be expressed from them with ease.

Cavernous hemangiomas occur alone or with a superimposed strawberry lesion. The cavernous elements are usually doughy in consistency and partially compressible and consist of capillaries and larger vessels, together with large vascular sinuses, all of which are located deeper than the vessels of the strawberry hemangiomas, thus imparting to the overlying tissues a bluish rather than red color. Strawberry hemangiomas are commonly found superimposed on cavernous lesions.

The *nevus flammeus* or "portwine mark" is made up of many telangiectatic vessels located quite superficially in the cutis and sometimes subcutis, thus causing the plaques to appear various shades of red and reddish-blue.

The natural history or clinical course of the majority of strawberry and cavernous hemangiomas is quite characteristic—spontaneous involution is the rule[13] and it usually takes place in the following sequence of events: The lesions are present at birth, or appear shortly thereafter. They then grow for several months, sometimes rapidly; but ordinarily they take from three to six months to reach their final dimensions. This is followed by a period of apparent quiescence when they begin to regress spontaneously. Complete disappearance is usually not achieved until the child is four, five or six years old. The clinical characteristics of lesions undergoing involution are rather typical and consist primarily of a lessening of the reddish color and an increase in the bluish tinge; the solid color is broken up by mottled grayish areas which represent the sites from which the vascular elements are disappearing; the tense, shiny surface becomes dull and wrinkles appear. These changes most frequently take place in the center of the lesion and then later involve the periphery. The resulting scar will depend on the original size and extent of the hemangioma and whether the involution was entirely spontaneous or followed ulceration of the surface of the lesion. In general, the final appearance of the residual scar is cosmetically and functionally more acceptable following spontaneous involution than following therapeutic intervention with solid carbon dioxide or, at times, the various other forms of therapy. Sufficient numbers of these hemangiomas have now been observed for spontaneous involution by various authors to substantiate the value of patience and avoidance of therapy in order to obtain the best cosmetic result. There are exceptions to this rule, however, as indicated below.

Many misconceptions regarding the management of vascular nevi exist. Only a few will be mentioned:

Misconception 1: All cavernous and strawberry hemangiomas disappear spontaneously; therefore, they do not require treatment.

As just stated, the majority of strawberry and cavernous hemangiomas do not require therapy because they can be expected to disappear spontaneously; there are, however, certain of these vascular tumors for which attention is necessary. These include: 1) rapidly enlarging hemangiomas; 2) hemangiomas encroaching on external orifices or vital organs; 3) hemangiomas involving wide spread areas; and 4) mucosal hemangiomas.

When the decision is made not to treat a hemangioma, the physician should explain to the parents the ordinary natural course of the majority of such lesions, pointing out, however, that therapy may become necessary. Parents and physician alike should be on the lookout for any unexpected changes, such as sudden enlargement, bleeding, ulceration, infection, necrosis, and interference with or destruction of adjacent vital structures. The patient, therefore, should be seen at regular intervals in order to re-examine the nevus and begin therapy if indicated.

Misconception 2: All strawberry and cavernous hemangiomas should be treated.

There are distinctly two schools of thought regarding the management of these tumors. Those who favor therapy argue that, because of the unpredictable course of hemangiomas, every lesion should be treated while it is still small, or as soon as it comes to medical attention, in order to prevent additional growth. Those, on the other hand, who advocate watchful waiting without treatment, do so by posing the questions, how many typical strawberry and cavernous hemangiomas are seen in adults? (Certainly very few indeed.) And how many unsightly scars or other sequellae are to be seen at the sites of spontaneous involution of these lesions? (Again, very, very few.)

And so the question whether to treat these nevi or leave them alone remains a constant challenge to the practicing physician, for it is *he* who is plagued by parents, grandparents, and other relatives to do something about "that unsightly growth". This is a responsibility that the physician should not take lightly. Should he subscribe to the teachings of the school which favors watchful waiting, as I do (unless there are distinct indications for therapy), he should have the opportunity to re-

examine the lesion at appropriate intervals, and each time should ask the following questions of himself[1]:

1. Does the hemangioma have an atypical appearance, or has its course of growth and involution been other than the usual one?

2. What permanent cosmetic defects may result without therapy, and how important will these defects be? Will therapy improve the chances for a better cosmetic result?

3. What impairment of function may there be or may there develop, if the hemangioma is left untreated (e.g., lesions of palms, soles, genitalia)?

4. What can therapy of the lesion be expected to accomplish that leaving it alone will not do? In other words, what would be the advantages of therapy?

Should therapy become indicated, the characteristics of the lesion, its location, size, depth, rate of growth, and all associated factors should be considered carefully in order to select the appropriate modality. Whatever method is decided upon should be used prudently and with full consideration, not only of therapeutic success or failure, but of the resulting cosmetic appearance.

Of the methods used for treating strawberry and cavernous hemangiomas, x-rays, radium, cryotherapy and the injection of sclerosing substances are the most frequently employed. While there may be specific indications for selecting one modality in preference to another, more often than not a particular method is chosen because it is the one best known to the physician, and/or the one with which he has had the most experience and in which he has the most confidence.

In general, we favor ionizing radiation when therapy is definitely indicated, provided it is given in sub-erythema doses and then only according to a schedule which allows for very slow involution of the nevus. In this way, where nature is given "a boost", so to speak, the cosmetic result is likely to simulate that achieved through spontaneous involution and the radiation dose remains well within limits of safety.

Because of the very superficial location of some of the smaller strawberry hemangiomas, solid carbon dioxide may sometimes be used to advantage. Treatment with this form of cryotherapy may be repeated every few weeks, with caution being taken not to produce excessive ulceration and a subsequent undesirable scar. Because of the superficiality of the freezing produced by carbon dioxide, this form of cryotherapy is of no value in the treatment of cavernous hemangiomas.

In addition, various sclerosing agents, as well as various surgical techniques, have been used successfully in the management of selected strawberry and cavernous hemangiomas.

Most important, therapy should be administered carefully and conservatively, for we have all seen the undesirable and unhappy results achieved through too vigorous cryotherapy, excessive doses of radiation and a haphazard injection of sclerosing substances. Even with surgical extirpation, recurrences have been known.

Misconception 3: There is no effective therapy for nevus flammeus.

With the knowledge that these lesions seldom fade appreciably and rarely disappear completely spontaneously, all too commonly the parent is told that nothing can be done for a child's birthmark when it is the ordinary, garden-variety portwine stain. Actually, there are three approaches which may be used:

1. The topical application of thorium X, a radioactive material which has been used in dermatologic practice for over 45 years without producing harmful sequellae. Treatments are given once every six weeks for as many as 10, 20 or more times[14]. In our experience, and that of others, about one third of the patients treated in this manner show considerable and satisfactory lightening of the lesion, another one third show but little improvement and the remaining one third show no response whatsoever.

2. Cosmetic camouflage. Certain of these lesions may be satisfactorily covered by special make-up preparations, such as pancake, "Spot-stik", and "Covermark" (Lydia O'Leary).

3. Also reported to be of value is the Conway technique of tattooing insoluble pigments into the cutis and subcutis above the vessels, thus hiding the lesion[15].

The proper management of these vascular nevi can be achieved only if the physician is familiar with their natural course, the complications which may occur, the various forms of therapy with their indications and contraindications, the therapeutic results that may be anticipated and the sequellae which may follow. He must be prepared to handle the unexpected emergencies and should seek consultative advice whenever needed.

For details concerning the diagnosis and therapy of these vascular nevi, here again, I must refer the reader to comprehensive texts and specific articles on the subject.

Summary and Conclusion

I shall conclude this very condensed presentation of a most important and broad subject by pointing out that 1) there is no substitute for experience in making the clinical diagnosis of pigmented lesions of the skin; 2) the therapy of nevus-cell nevi and hemangiomas must be selected to meet the needs of the particular situation, and lastly, 3) in the management of skin lesions the therapeutic modality should be carefully chosen so as to achieve not only a rapid and permanent cure but, whenever possible, one that will be cosmetically and functionally acceptable.

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